

Surgeons refine procedure for life-threatening congenital heart defect

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For children born with life-threatening hypoplastic left heart syndrome (HLHS), reconstructive surgeries can restore blood circulation. While the most common corrective approach is the three-stage Norwood procedure, an alternative strategy, hybrid palliation, allows deferral of the more complex reconstructions to when the child is somewhat older and better able to successfully recover from major surgery. A report in *The Journal of Thoracic and Cardiovascular Surgery*, the official publication of the American Association for Thoracic Surgery (AATS), evaluates whether an arterial shunt in the hybrid palliation may be a better source for the pulmonary blood supply than the more frequently used venous shunt.

Each year, almost 1000 babies in the United States are born with HLHS, a congenital condition in which the left side of the heart is undeveloped and systemic blood flow is inadequate to sustain life. Without surgical intervention, either reconstruction of structures of the heart and blood vessels or cardiac transplantation, HLHS is fatal. Symptoms of HLHS manifest hours or days after birth, when the ductus arteriosus, a blood vessel connecting the [pulmonary artery](#) and the aorta, begins to close. Symptoms include breathing problems, pounding heart, weak pulse, ashen or bluish skin, and heart murmurs. The three-stage Norwood procedure occurs at different times of development, with Norwood Stage I typically performed soon after birth, Norwood II (commonly referred to as a bidirectional Glenn - or cavopulmonary - shunt) between 4 and 6 months of age, and the final surgery, termed a modified Fontan Procedure, between 2 and 5 years of age.

Although Norwood palliation has achieved 30-day survival rates of 90% or more, surgeons strive for better ways to improve outcomes for the highest risk patients. "Hybrid palliation was initially thought to be a therapy that would eventually supplant standard Norwood palliation because of its technical simplicity, its avoidance of cardiopulmonary bypass (open heart surgery) and prolonged perioperative recovery in the neonatal period, and an intuitive notion that it would be associated with improved neurodevelopmental outcomes," commented David M. Overman, MD, Chief of the Division of Cardiovascular Surgery at the Children's Hospitals and Clinics of Minnesota (Minneapolis) in an accompanying editorial.

"In our center, the hybrid procedure is reserved for higher-risk, more complex, and unstable patients in whom a traditional Norwood procedure would carry an unacceptably high risk," explained lead investigator Mohamed S. Nassar, PhD, FRCS, of the departments of Paediatric Cardiology and Cardiac Surgery, Evelina London Children's Hospital, Guy's and St. Thomas' NHS Foundation Trust (London).

In the second stage of the hybrid strategy, the aortic arch must be reconstructed and a source of blood supply to the lungs must be established. In a quest to refine the hybrid procedure, surgeons may use two possible sources for the pulmonary blood supply. The usual practice was to create a venous shunt between the pulmonary artery and superior vena cava (a cavopulmonary shunt). However, other investigators have raised concerns about a venous shunt, preferring instead an initial arterial shunt constructed between the first branch of the aortic arch and the right pulmonary artery (modified Blalock-Taussig shunt), followed later with the superior cavopulmonary anastomosis.

"The clinical issue driving this novel strategy is the increasingly well documented problem of branch pulmonary artery stenosis associated with hybrid palliation of HLHS," noted Dr. Overman. "The authors'

premise that use of an arterial shunt at the time of arch reconstruction may result in improved pulmonary artery architecture is reasonable."

Since both the arterial and venous shunts were being performed at the same institution, the researchers had an ideal opportunity to conduct a retrospective analysis of their cases to see whether one procedure offered better outcomes than the other. Dr. Nassar identified 17 HLHS patients who received an arterial shunt and 26 patients who received a venous shunt. Indeed, the arterial group did show better pulmonary arterial growth than the venous shunt, as indicated by a higher lower lobe index.

There are other pros and cons to each procedure. Surgical times were shorter in the arterial group, but this group also demonstrated a higher need for delayed sternal closure. Mechanical ventilation and intensive care stay were shorter in the venous shunt group. At discharge, there was no difference in the echocardiography findings, but those who received arterial shunts had significantly higher oxygen saturations.

No differences were found in mortality or incidence of complications between the two approaches. Both groups had approximately a 30% rate of MRI-proven brain abnormalities after clinically detected abnormal neurological findings in the immediate postoperative period. "This relatively high incidence is comparable to other reports studying the neurologic development in patients with HLHS," stated Dr. Nassar.

Dr. Overman expressed his concern with the high rate of clinically detectable postoperative neurologic abnormalities and associated MRI findings in both treatment groups. "The central tenet of hybrid strategy is that neurodevelopmental outcomes will be improved by delaying larger reconstructive operations from the neonatal period into infancy."

While Dr. Overman noted that hybrid surgery strategies for HLHS are

currently used only for a minority of patients, with the Norwood procedure still being preferred at most institutions, he acknowledged that there is a place for hybrid surgery in a higher-risk subset of patients.

"The impact and advisability of that particular approach, while intuitively resonant, is still an open question. The arterial shunt at Stage II is yet another twist in the evolving story of hybrid therapy for HLHS." Dr. Nassar and co-investigators found that age and weight at second stage were lower in the arterial group than the venous group, suggesting that patients who had undergone the arterial shunt included those at higher risk. What is clear is that surgery for HLHS is an ongoing story as surgeons seek better outcomes with fewer complications for babies born with this serious abnormality.

Provided by American Association for Thoracic Surgery

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